

Nutritional Disorders

Adequate Diet

It should fulfill the following criteria:

1. Adequate total calories.
2. Adequate proportions of protein, fat & CHO.
3. Adequate amount of vitamins & minerals.
4. Adequate amount of water.

Balanced diet

It should contain adequate proportion of fat, CHO & proteins:

1. CHO $\Rightarrow$ 50 % (1 gm = 4.1 Cal).
2. Fat $\Rightarrow$ 35 % (the main source of energy , 1 gm = 9.3 Cal).
3. Proteins $\Rightarrow$ 15 % (1 gm = 4.1 Cal)

How to calculate the caloric requirement for normal child ?

Premature: 120 Cal / Kg / day.

Newborn: 100-120 Cal / Kg / day.

Children:

- 1st 10 Kg $\Rightarrow$ 100 Cal / Kg / day.
- 2nd 10 Kg $\Rightarrow$ 50 Cal / Kg / day.
- Over 20 Kg $\Rightarrow$ add 20 Cal / Kg / day.

Ideal diet

1. Adequate diet.
2. Palatable.
3. Available.
4. Cheap.



Come on .. Do your best

Abnormal Nutrition

Under-nutrition

Due to deficiency in the total caloric supply as well as deficiency in the 3 basic food stuffs (CHO - fat - proteins) & deficiency of minerals & vitamins e.g. marasmus.

Malnutrition

Deficiency or excess of one or more of the food stuffs with almost normal total caloric supply e.g.

1. ↓↓↓ Vitamin D $\Rightarrow$ Rickets.
2. ↓↓↓ Proteins $\Rightarrow$ KWO.
3. ↓↓↓ Fe $\Rightarrow$ iron deficiency anemia.
4. ↑↑↑ Calories $\Rightarrow$ obesity.
5. Hypervitaminosis.

Mixed malnutrition & under-nutrition

There is deficiency of one or more of food stuffs + deficiency of total caloric supply e.g. marasmic KWO.

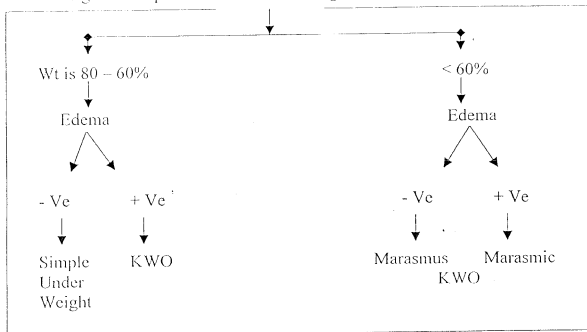


Work hard .. It's great challenge

Classification of Nutritional Disorders

Welleome Classification 1970

Protein - Energy Malnutrition " PEM" is classified according to the weight of the patient in relation to its age as follows



Marasmus "Infantile Atrophy"

Definition

It is a state of chronic under-nutrition in the form of deficiency of total caloric supply as well as deficiency of different food stuffs. It is characterized by progressive loss of weight reaching below 60 % of the normal.

Etiology

I. Dietetic Errors (1ry marasmus):

A. Quantity

1. Scanty breast milk.
2. Small amount of feed
3. Widely spaced feeds
4. Delayed weaning.

B. Quality

1. Diluted formula in artificial feeding.
2. Cow-milk protein allergy.

C. Nursing Errors

1. Irregular feeding
2. Air-swallowing with failure of eructation.

D. Feeding difficulties:

1. *Breast fed*: in both baby & mother.
2. *Artificial fed*: in the baby only.

E. In weaned infant:

Failure to maintain well balanced diet.

II. Non dietary causes (2ry marasmus)

1. Infections:

- GE (the commonest) only if prolonged or recurrent.
- TB.
- Chronic chest infections.
- Chronic suppurative otitis media.
- Urinary tract infection.
- Congenital infections.

2. GIT causes:

a. Congenital:

- Cleft lip & palate.
- Congenital hypertrophic pyloric stenosis.

b. Malabsorption syndromes

3. Chronic systemic disorders:

- a. Congenital heart diseases.
- b. Recurrent or intractable heart failure.
- c. Renal tubular acidosis.
- d. Obstructive uropathy.
- e. Chronic renal failure.
- f. Congenital lung cysts.
- g. Hepatic cirrhosis.
- h. Severe mental retardation.
- i. Neurological deficits e.g. palatal paralysis.

4. Prematurity

Due to

1. Weak suckling
2. Poor absorption
3. Poor stores
4. High caloric needs/Kg

5. Metabolic Diseases

1. Glycogen storage disease.
2. Galactosemia.
3. Phenylketonuria.



6. Endocrine Diseases

1. Thyrotoxicosis.
2. D.M.

7. Chromosomal disorders.

Pathology

Defective total caloric intake $\Rightarrow$ utilization of spare tissues (hepatic glycogen & fat) for energy production

Persistence of the condition $\Rightarrow$ utilization of fixed tissues (proteins in the muscles & organs) $\Rightarrow$ Picture of generalized atrophy

1. Loss of hepatic glycogen.
2. Loss of sub-cutaneous fat
3. Atrophy of muscles & other internal organs
4. Bones $\Rightarrow$ Osteoporotic "Atrophic-rickets"

C/P

C/O

The usual C/O is failure to thrive

But it may be presented by one of the complications or etiological factors.

History

1. History of growth delay. "developmental history"
2. History of nutritional troubles "Dietetic history"
3. History of one of the etiological factors or complications.

O/E

1. Growth failure

- First there's failure to thrive $\Rightarrow$ Followed by progressive loss of wt.
- This can be detected early from the growth curve of the baby.
- According to wellcome classification the wt is less than 60% of expected.

2. Loss of S.C. Fat

- There's marked loss of S.C. fat $\Rightarrow$ $\downarrow$ Skin fold thickness.
- The skin becomes thin & wrinkled esp. at the flexural areas.

- According to its distribution marasmus can be

Senile face
"3rd degree"

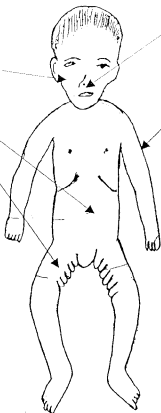
Abdominal Wall
"1st degree"

Limbs & buttocks
"2nd degree"

3. Muscle Wasting

marked prominence of bony surfaces

The whole picture is that of skin over bone



4. Marked pallor

5. Mid-arm circumference

The marked atrophy of ms. & S.C. Fat $\Rightarrow$ $\downarrow\downarrow$ midarm circumference

N.B.

- Normally at the age of 1 year $\Rightarrow$ > 13.5 cm

- In early cases = $12.5 \div 13.5$ cm

- In full marasmic pts = < 12.5 cm

6. Skin manifestations:

- Loss of skin elasticity.
- Skin is thrown into multiple folds especially at the groins.
- So we can not depend on loss of skin elasticity as a sign of dehydration in marasmus

7. Abdomen:

- Scaphoid abdomen due to thin abdominal muscles.
- Visible peristalsis due to thin abdominal muscles.

8. Irritability: due to hunger pains.

9. Hypothermia: due to :-

- a. $\downarrow\downarrow\downarrow$ Caloric intake.
- b. Loss of S.C. fat $\Rightarrow$ increase heat loss.
- c. $\downarrow\downarrow\downarrow$ Muscle bulk $\Rightarrow$ impaired shivering.
- d. Dehydration & electrolyte imbalance.
- e. Septicemic shock.

10. GIT manifestations

- The patient is always hungry, but anorexia is present in advanced cases.
- Constipation $\Rightarrow$ may be present due to decrease food intake.
- Diarrhea $\Rightarrow$ due to

1. G.E.
2. Maldigestion
3. Malabsorption (commonly lactose intolerance).
4. Starvation diarrhea.

N.B.

Starvation diarrhea is common

• Characters

1. Small amounts
2. Loose
3. Dark in color
4. Offensive odor

• It is composed of

1. Cellular debris.
2. Greenish mucus.
3. Intestinal flora.

11. CVS:-

- Weak & slow pulse.
- In case of dehydration $\Rightarrow$ weak & rapid pulse.
- Peripheral circulatory failure is the end stage.

Marasmus

12. Respiratory system:-

- Shallow respiration $\Rightarrow$ due to weak respiratory muscles.
- Repeated chest infection.

13. Signs of mineral & vitamin deficiency $\Rightarrow$ see later.

14. Inter-current infections (due to impaired immunity)

1. G.E.
2. Bronchopneumonia "Main cause of death"
3. O.M.
4. Urinary tract infections
5. T.B.

Investigations

Marasmus is a clinical diagnosis, but we need investigations for diagnosis of the cause & complications if present.

1. Stools:

- Parasites
- Steatorrhea

- Malabsorption.

2. Urine:

- Culture & Sensitivity
- Sugar & Ketones $\Rightarrow$ DM
- $\uparrow\uparrow$ Urea due to hyper-catabolism of proteins.

3. CBC

- a. Anemia $\Rightarrow$ see KWO later.
- b. $\uparrow\uparrow\uparrow$ Total & differential WBCs due to infection.
- c. $\uparrow\uparrow\uparrow$ ESR due to infection.

4. Chemistry:

- a. Slight hypoproteinemia.
- b. $\uparrow\uparrow\uparrow$ Igs due to infections.
- c. Fasting hypoglycemia.
- d. Serum electrolytes may be disturbed due to GE & dehydration.

5. CXR

- Congenital heart disease
- T.B.
- Bronchopneumonia.

6. Tuberculin test $\Rightarrow$ For T.B.

7. Intestinal biopsy.

8. Estimation of digestive enzymes

Complications

1. Intercurrent infections

2. Hemorrhagic tendency & purpura It is a bad sign.

It is due to:

- a. Loss of SC support of blood vessels.
- b. DIC due to severe dehydration & fulminant infection.

3. Edema $\Rightarrow$ Marasmic KWO if the patient is fed on CHO diet with deficient protein.

4. Hypothermia

5. Hypoglycemia due to lack of glycogen stores in the liver.

6. Dehydration & electrolyte imbalance, acid base disturbances & shock.

7. Peptic ulceration.

Kwashiorkor KWO

Definition

It is a clinical syndrome which results from malnutrition in the form of deficient protein intake with adequate total caloric intake through a CHO diet.

- So, it is called **Protein Energy Malnutrition**

N.B.

° It is also called the disease of deposed baby because it usually affects the elder baby when the next is born (i.e. Maternal deprivation).

Epidemiology

- Mostly at the age of 6 ms -3 Years following & coincident with weaning
- Rare < 6 months.
- Never > 3 years because he can struggle for eating.
- Common in low socio-economic class.

Etiology

• Primary KWO (Dietetic)

- Unbalanced diet, with minimal protein intake & high CHO intake.
- This is common to occur at the time of weaning.
- Also, faulty management of marasmic babies, by giving them a CHO diet only will result in marasmic-KWO.

• Secondary KWO:

- Resulting from other diseases, which are usually predisposing factors rather than actual causes as these diseases will lead to:
 1. Anorexia & stomatitis.
 2. Malabsorption & protein losing enteropathy.
 3. Prolonged dietary restriction e.g.
 - a. GE.
 - b. Whooping cough.
 - c. Measles.
 - d. Parasitic infestation.

<i>Essential Features</i>	<i>Variable features</i>
1. Growth retardation 2. Edema 3. Mental changes 4. $\downarrow\downarrow$ Ms / fat ratio	1. Hair changes 2. Skin changes 3. Hepatomegaly 4. GIT troubles 5. Vitamins & mineral deficiency 6. Anemia 7. Infections

I. Growth retardation

- Early, there is slow rate of growth when measured on growth curves.
- The wt is affected $>$ height
- The wt is usually 60-80% of the expected.
- The loss of weight may be masked by edema & excess SC fat.
- In advanced cases bone age may be delayed.

II. Edema

It is characterized by

1. Pitting
2. Site: starts in the dorsum of the hands & feet then spreads centrally, to involve legs, arms, face & does not affect the anterior abdominal wall.
3. It never causes ascitis or effusion.

Causes:

1. Low serum albumin $\Rightarrow$ is the main cause $\Rightarrow \downarrow\downarrow$ effective plasma oncotic pressure.
2. 2ry Hyper-aldosteronism
 $\Rightarrow$ Why?
3. $\uparrow\uparrow$ ADH level $\Rightarrow$ Why?
4. Altered capillary permeability due to protein deficiency.

III. Mental Changes

These are represented by lethargy, apathy, misery, Sluggish movement, anorexia & disinterest in the surroundings.

Causes

$\Rightarrow$ Causes of brain pathology.

Pathology

I. Fatty liver $\Rightarrow$ Hepatomegaly

It never leads to cirrhosis except if the condition is prolonged.

Causes

1. Deficiency of lipotropic factors e.g. Methionine & Choline,
2. Defective formation of lipoproteins $\Rightarrow$ due to $\downarrow\downarrow$ plasma proteins.,
3. Vit- B complex deficiency.

II. Excess S.C. Fat

III. Atrophy of intestinal villi.

- $\Rightarrow$ 1. Malabsorption
2. $\downarrow\downarrow$ Disaccharidase enzymes $\Rightarrow$ Mal-digestion (commonly lactase deficiency $\Rightarrow$ lactose intolerance).

IV. Generalized muscle wasting

$\Rightarrow$ due to $\downarrow\downarrow$ protein

V. Pancreas

Atrophy of pancreatic acini

$\Rightarrow \downarrow\downarrow$ pancreatic enzymes

$\Rightarrow$ Malabsorption & maldigestion

VI. Heart

Degenerative changes $\Rightarrow$ H.F.

VII. Kidney $\Rightarrow$ Hyaline degeneration of the glomeruli & may be tubular cells.

VIII. Brain

Slow atrophy $\Rightarrow$ Mental changes

Causes

1. $\downarrow\downarrow$ Aromatic A.A. tryptophan tyrosine & phenyl -alanine which are needed for proper growth of the brain.
2. Hypothyroid function

N.B.

• Mental changes are reversible by t t t. however if the condition is severe & left without ttt in young age $\Rightarrow$ permanent mental affection.



IV. ↓↓ Muscle / Fat ratio

1. The muscles are markedly atrophied this atrophy is best palpated on the biceps, triceps, deltoid or quadriceps muscles.
2. At the same time S.C. fat is preserved or even increased due to adequate or high total caloric intake $\Rightarrow$ CHO facies (moon face baby).

V. Skin Manifestations

- Skin usually shows erythema & hyper-pigmentation, followed by desquamation, hypopigmentation & ulceration.
- Fissuring & cracking of the skin may give the picture of flaky paint.
- 2ry infection & even gangrene of the skin may occur. $\rightarrow$ Why?
- Napkin dermatitis is common & usually complicated by fungal & bacterial infection of the skin.
- Sites $\Rightarrow$ it can occur all over but the commonest sites are.
 1. Pressure area $\Rightarrow$ buttocks & back
 2. Flexural areas $\Rightarrow$ groin & axillae
- Causes
 1. ↓↓ Essential F.A.
 2. ↓↓ Nicotinamide & tryptophan
 3. ↓↓ Vit. A.
 4. ↓↓ Zinc

VI. Hair Changes

- The hair loses its luster, becomes easily pickable & sparse.
- The colour changes into dark brown $\Rightarrow$ light brown $\Rightarrow$ red $\Rightarrow$ yellow $\Rightarrow$ white.
- Flag sign $\Rightarrow$ the distal areas of hair are darker than the proximal areas.
 - Causes
 1. ↓↓ Sulfer containing A.A. methionine & cystin.
 2. ↓↓ Copper & Ceruloplasmin
 3. ↓↓ Melanin formation.

VII. GIT Disturbance

- Anorexia & vomiting due to:
 - a. Mental changes.
 - b. Associated GIT infection.

- *Diarrhea* $\Rightarrow$ which may be due to
 1. 2ry Disaccharidase deficiency e.g. Lactose intolerance
 2. 2ry Infections "GE"
 3. $\downarrow\downarrow$ Digestive enzymes "Pancreatic & intestinal"
 4. Starvation diarrhea.
 5. $\downarrow\downarrow$ Fe $\Rightarrow$ atrophy of the epithelium.

VIII. Anemia

Which may be due to one of the following:

1. $\downarrow\downarrow$ Iron or Cu $\Rightarrow$ Microcytic hypochromic
2. $\downarrow\downarrow$ B₁₂ & Folic acid $\Rightarrow$ Megaloblastic.
3. $\downarrow\downarrow$ Protein $\Rightarrow$ Normocytic normochromic
4. Infection $\Rightarrow$ $\ominus$ BMor hemolysis $\Rightarrow$ Normocytic normochromic.

IX. Abdominal distention :

1. Hypokalemia (2ry to diarrhea).
2. Weak abdominal muscle.
3. Infection $\Rightarrow$ toxic ileus.
4. Malabsorption $\Rightarrow$ fermentation $\Rightarrow$ gaseous distention.
5. Giardia infection.

X. Vitamins & Minerals Deficiency

A. Vitamin A : It is very common in KWO due to :

1. $\downarrow\downarrow$ intake in diet.
2. $\downarrow\downarrow$ Storage of carotene inside the liver.
3. $\downarrow\downarrow$ Conversion carotene to vitamin A inside the liver due to Zn deficiency.
4. $\downarrow\downarrow$ Carrier proteins.

Clinical manifestationssee later in vitamin deficiency.

B. Vitamin B2: Angular stomatitis, glossitis & vascularization of the cornea.

C. Vitamin D: Atrophic rickets.

D. Nicotinic acid: Pellagra.

E. Vitamin C: Scurvy.

F. Cu: Anemia, hair changes & mental changes.

G. Zn: Mental, hair & skin changes.

XI. Associated infections:

It occurs due to defective immune system (chemotaxis, intracellular killing, complement system & opsonization).

Investigations

A. Sub-Clinical Cases

1. Serum Albumin: $< 2.85 \text{ gm \%}$ but $> 2.5 \text{ gm \%}$
2. Serum Amino Acids:
 - Low serum A.A.
 - The ratio of Non-essential/ Essential
 - a. Normally = 2/1
 - b. Early cases = 2/1 – 3/1
 - c. Clinical cases = $> 3/1$

B. Clinical Cases

1. Serum proteins
 - a. $\downarrow\downarrow$ Total proteins
 - b. $\downarrow\downarrow$ S. Albumin $< 2.5 \text{ gm/dl}$
 - c. $\downarrow\downarrow$ α , β globulins, while γ globulin may be $\uparrow\uparrow$ due to high incidence of infections.

N.B.

- Globulin is formed in the RES & severe protein deficiency is needed to cause failure of the RES.
- There is thymic atrophy $\Rightarrow \downarrow\downarrow$ T-helper cells.

2. Serum amino acids $\Rightarrow$ see before.

3. $\downarrow\downarrow$ Carrier proteins e.g. Ceruloplasmin – transferrin – Thyroid binding globulin.

4. All body enzymes are decreased except

- a. SGOT(AST) $\Rightarrow \uparrow\uparrow$ due to hepatocytic damage & muscle damage.
- b. SGPT(ALT) $\Rightarrow \uparrow\uparrow$ due to hepatocytic damage

4. Disturbed fat metabolism:

- a. $\uparrow\uparrow$ Free fatty acids.
- b. $\downarrow\downarrow$ Serum cholesterol due to:
 - $\downarrow\downarrow$ Carrier proteins.
 - $\downarrow\downarrow$ Lipotropic factors.

5. Hypoglycemia

⇒ There's fasting hypoglycemia with impaired glucose tolerance curve.

6. CBC

⇒ Anemia of many types + ↑↑ ESR & WBCs.

⇒ "See before"

7. Urine

⇒ for UTI + ↓↓ urinary urea & hydroxy proline in urine.

8. Stools

⇒ as marasmus

8. CXR & Tuberculin test

9. Water & electrolytes:

- Na: ↑↑ total body Na but there is dilutional hyponatremia.
- K : ↓↓ due to diarrhea & hyperaldosteronism.
- ↓↓ Cu, Mg, Zn & P.
- ↓↓ Total serum Ca (↓↓ protein bound form & normal ionizable form) ⇒ no tetany.

D.D.

<i>Of edema</i>	<i>Of skin – changes</i>
1. Renal ⇒ Nephritic Nephrotic	1. Pellagra
2. Cardiac	2. Napkin dermatitis
3. Hepatic	3. Acrodermatitis enterop- Athica
4. Allergic ⇒ Angio-neurotic	4. Hartnup disease.

Napkin dermatitis

It is due to:

1. Amoniacal dermatitis: The most common cause

Mechanism ⇒ continuous soiling of the infant by his urine ⇒ bacterial splitting of urea ⇒ production of ammonia ⇒ irritation & excoriation of the skin.

2. Prolonged soiling: $\Rightarrow$ humidity $\Rightarrow$ excoriations.

3. Monifial infection: red, moist lesion with well defined raised edges. There is characteristic satellites outside the margin.

4. Over-feeding.

5. Rough clothes $\Rightarrow$ continuous friction.

6. Acidic diarrhea e.g. lactose intolerance & GE.

7. Contact dermatitis: due to polysynthetic materials or application of chemicals.

Infantile pellagra

Occurs in sun exposed areas & over bony prominences.

Hartnup disease

Autosomal recessive disease characterized by defective intestinal transport of tryptophan $\Rightarrow$ Niacin deficiency $\Rightarrow$ pellagra like skin manifestations.

Acrodermatitis enteropathica

Autosomal recessive disease characterized by defective intestinal absorption of Zn.

C/P:

1. Chronic diarrhea.
2. Skin lesion as in KWO especially in perianal area.
3. Nail dystrophy.
4. Stomatitis.
5. Growth retardation.
6. Intercurrent infection.

Complications

1. Inter-current infections

2. G.E. $\Rightarrow$ dehydration & electrolyte imbalance

3. Hypoglycemia $\Rightarrow$ Why?

- $\downarrow\downarrow$ Glycogen stores inside the liver.
- $\downarrow\downarrow$ Counter-regulatory hormones of insulin e.g. catecholamines & glucagon.

4. Hypothermia $\Rightarrow$ Why?

- Severe dehydration & shock.
- Septicemic Shock.
- Impaired shivering due to muscle wasting.
- Hypoglycemic attacks.

5. DIC (bad prognostic sign) $\Rightarrow$ Why?

6. Metabolic complications:

- $\downarrow\downarrow$ Ca, Mg, Na, K.

7. HF $\Rightarrow$ Why?

- Degenerative changes in cardiac muscles.
- Anemic HF.
- Toxic myocarditis.
- Volume overload.

Marasmic - KWO

- Baby with all essential features of KWO + Wt $< 60\%$ of normal wt for his age.
- It is due to combination of:
 1. $\downarrow\downarrow$ Total caloric intake "under-nutrition"
 2. $\downarrow\downarrow$ protein "mal-nutrition"
- C/P:
 - Loss of SC fat.
 - Lack of moon face of KWO.
 - With or without skin & hair changes .
- !!! is more difficult & prognosis is bad

Pre-KWO " CHO- baby

- Baby with incomplete picture of KWO " Absence of one or more of the essential features"
- There is decrease body weight.
- Mental changes.
- Hair & may be skin changes.
- No edema.

Nutritional Dwarfism

In this condition, early mild to moderate protein-energy undernutrition occurs, to which adapts himself by slow growth $\Rightarrow \downarrow \downarrow$ both Wt & height " $< 10^{\text{th}}$ percentile for age" $\Rightarrow$ Proportionate dwarfism.

So, in this condition:

1. Wt for age $\Rightarrow$ low " $< 10^{\text{th}}$ percentile"
2. Height for age $\Rightarrow$ Low " $< 10^{\text{th}}$ percentile"
3. Wt for height $\Rightarrow$ normal proportionate"

Causes of Hypoproteinemia

1. $\downarrow \downarrow \downarrow$ Intake
2. Indigestion & Malabsorption
 - a. Chronic diarrhea
 - b. Celiac disease
 - c. Intestinal parasites
3. $\downarrow \downarrow \downarrow$ Loss
 - a. Urinary $\Rightarrow$ Nephrotic syndrome
 - b. Intestinal $\Rightarrow$ Protein losing enteropathy.
 - c. Cutaneous $\Rightarrow$ Burns, Ulcers, Eczema.
 - d. Others $\Rightarrow$ Repeated taps for ascitis, hydrothorax.
4. Decreased Production of serum proteins "Albumin"
 - a. Liver diseases.
 - b. Inborn errors of albumin.
e.g. Analbuminemia.

Management of Nutritional Diseases

1. Proper history
2. Proper Clinical examination
3. Investigations.
4. D.D.
5. Treatment.



Just do your best

Treatment of Marasmus & KWO

A. Prevention

1. Proper diet
2. Proper breast feeding.
3. Proper weaning
4. Proper vaccination esp. against measles, whooping cough, T.B.
5. Family planning.
6. Follow up of the growth of the baby by " growth curves $\Rightarrow$ for early detection & ttt.

B. Dietetic Management

1. Education of the mother about the nutritive needs of her baby & how to prepare adequate diet is essential.
2. Caloric intake / Day.
 - a. In marasmus $\Rightarrow$ 150 Cal./Kg/D
 - b. In KWO $\Rightarrow$ 120 Cal./kg/D

In 1st degree marasmus it is calculated according to the expected weight, while in 2nd & 3rd degree marasmus & in KWO it is calculated on the average weight (mid-way between expected & actual body weight).

3. The diet should supply high protein "4 gm /Kg/D" of high biological value esp. in KWO.
4. Route of administration:
 - a. Oral : By bottle, cup & spoon or dropper.
 - b. Nasogastric tube: If there's vomiting , anorexia, severe stomatitis or cleft lip & palate.
 - c. Total parenteral nutrition: if there is contraindication of oral feeding or persistent vomiting inspite of nasogastric feeding..

* *In weaned babies* $\Rightarrow$ Yogurt, Vegetable soup, cereals, minced meat. Then give well balanced diet..

** *In non weaned babies* $\Rightarrow$

1. In uncomplicated cases:
Humanized milk + protein added

N.B.

2. In lactose intolerance:

Lactose free milk + protein

3. Fat intolerance

Skimmed milk + medium chain triglyceride + protein.

The protein added to the formula should be of high biological value.

- The amount & concentration of the formula should be increased gradually according to the tolerance of the baby, till the expected weight is reached.

C. Non-Dietetic Management

I. Emergency Management

In complicated cases the following should be done on emergency basis:

1. Shock

⇒ Stimulants & blood transfusion.

2. Dehydration

⇒ Rehydration & correction of electrolyte imbalance.

3. Hypothermia

⇒ Proper warming of the baby

4. Hypoglycemia

⇒ Glucose 10% by IV drip.

II. Blood transfusion + I.V. alimentation

1. Fresh blood ⇒ 20ml / Kg (the best)

2. FFP ⇒ 10ml/kg

3. Salt free albumin "Human" ⇒ 10ml/kg.

4. Amino-acid mixture preparation " Trophysan - Totamine - Vamine": 20 ml / Kg. ⇒ May cause hypoglycemia as they are rich in arginine a.a. ⇒ stimulation of insulin release.

It is contraindicated in HF, renal failure & hepatic failure.

N.B.

The advantages of blood transfusion are :

1. Correction of anemia.
2. ↑↑ Resistance to infection.
3. ↓↓ Edema in KWO.
4. Improvement of appetite.
5. Plasma proteins can be used for formation of the important critical proteins of the body e.g. enzymes hormones etc.
6. Correction of bleeding tendency fresh blood.

III. Antibiotics

In marasmus & KWO infections are common, either manifest or hidden.

⇒ Procaine penicillin or crystallin penicillin.

In Manifest infections ⇒ Culture & sensitivity should determine the appropriate antibiotic.

If there is moniliasis:

- Oral moniliasis ⇒ Mycostatin drops & gentian violet paint.
- Oral moniliasis ⇒ Nystatin cream & gentian violet paint.

5 Km

IV. Vitamins & Minerals

These should be supplied orally from the start. If diarrhea is present, we give them by IMI.

- Vit. A in high doses (50.000 IU / D) is particularly important if there is eye affection to prevent further damage & blindness.
- Zinc & magnesium should be supplemented especially in KWO.
- Oral Ca, or IV in tetany.

V. Anabolic Hormones

As Durabolin, Verabolin 1mg/Kg/D in order to help a positive nitrogen balance. However, their role in t t t is questioned.

VI. t t t of the cause

This is an essential part in management.

Q: What are the phenomena occurring during t t t of KWO ?

1. Over-Hydration during rehydration.
2. Hypokalemia ⇒ usually present at the start. Failure of diagnosis lead to death.
3. Nutritional recovery syndrome:

Which is noticed in nutritionally deficient babies, who are treated by high protein diets as in case of KWO.

It is caused by hepatic dysfunction in cases of KWO + high protein diet.

It presents by

- a. Hepatomegaly
- b. Hyper-ammonemia
- c. Encephalopathy with or without convulsions.

Usually, there are changes in the mood, behavior & level of consciousness in severe cases.

- d. Mild jaundice may be present.

Prognosis of KWO

1. Early : depends on :

- a. Age: young age $\Rightarrow$ bad prognosis.
- b. Cause : irreversible cause $\Rightarrow$ bad prognosis .
- c. Presence of complication $\Rightarrow$ bad prognosis.
- D. ttt : early ttt $\Rightarrow$ good prognosis

2. Later :

- a. Growth $\Rightarrow$ early sufficient ttt $\Rightarrow$ normal growth.
- b. Mentality $\Rightarrow$ MR occurs in early cases before 6ms or in sever cases.
- c. Liver cirrhosis $\Rightarrow$ does not occur.



Calcium Metabolism

Factors Affecting Absorption

1. PH of Duodenum

An acidic PH of the duodenum $\Rightarrow \uparrow\uparrow\uparrow$ Absorption.

Alkaline PH $\Rightarrow \downarrow\downarrow\downarrow$ Absorption.

2. Phosphate in Diet

If $\uparrow\uparrow\uparrow \Rightarrow$ precipitation of $\text{Ca}^{++} \Rightarrow \downarrow\downarrow\downarrow$ absorption.

N.B.

The Ca: P ratio in diet must be 2 : 1 or 1 : 1 for optimum absorption.

"This is the ratio present in human milk"

3. Other constituents of diet

A. Phytate "in cereal grains" $\Rightarrow \downarrow\downarrow\downarrow$ absorption

B. Oxalates "e.g. in spinach" $\Rightarrow \downarrow\downarrow$ absorption

C. High protein & vitamin C in diet $\Rightarrow \uparrow\uparrow$ absorption.

4. Hormonal factors

A. Vitamin D $\Rightarrow \uparrow\uparrow\uparrow$ absorption

B. Parathyroid hormone $\Rightarrow \oplus$ Formation of active form of vit - D
 $\Rightarrow \uparrow\uparrow$ absorption.

Blood Forms

- The normal plasma level is 9-11 mg%
- It is present mainly in 2 main forms:

A. Ionized form

$\Rightarrow$ The physiological active form

B. Bound form

$\Rightarrow$ Inactive form which is bound to plasma proteins

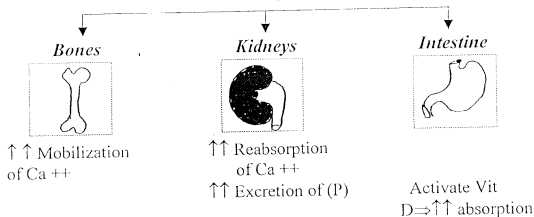
Concentraaaaaate ..

Factors Affecting Blood Ca^{++}

A. Para-thyroid hormone "PTH"

$\Rightarrow \uparrow \uparrow \uparrow \text{Ca}^{++}$
 $\downarrow \downarrow \downarrow (\text{P})$

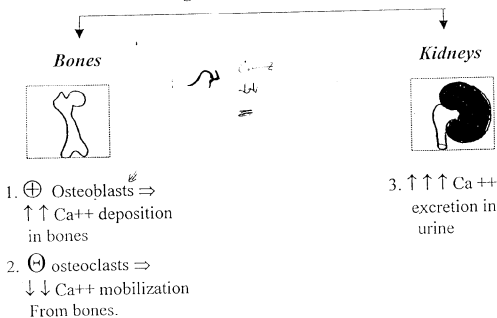
Through the following mechanisms



B. Calcitonin

$\Rightarrow \downarrow \downarrow \downarrow \text{Ca}^{++}$

Through the following mechanisms



Success wants more efforts

C. Vit - D

Bones



Deposition of Ca^{++}
& P

Kidneys



Reabsorption
of P & Ca^{2+}

Intestine



Absorption
of Ca^{++} & P

D. PH of the blood

Acidosis $\Rightarrow \uparrow \uparrow$ Ionizable form

Alkalosis $\Rightarrow \downarrow \downarrow$ Ionizable form



*Do you want to be one of
the fronts*

Rickets

Definition

It is a systemic metabolic disease characterized by defective mineralization of bone & cartilage in the growing baby, leading to excessive formation of osteoid tissue, which gives way under pressure leading to deformities.

Classification

A. Vit-D Deficiency Rickets

1. Dietary deficiency of vit-D
2. Lack of exposure to UVR
3. Atrophic rickets.

B. Vit-D Resistant Rickets

I. Renal Rickets:

1. Glomerular $\Rightarrow$ Renal osteodystrophy
2. Tubular $\Rightarrow$ 1ry hypophosphatemic Rickets
Lignac syndrome
De Toni-Fanconi Syndrome
Lowe's syndrome
Infantile renal tubular acidosis.

3. Vit-D Dependent Rickets

Typ I
Type II

II. Hepatic Rickets

1. Defective 25-hydroxylation
2. Toxic hepatic hydroxylation.
3. Malabsorption of Vit-D due to bile deficiency.

III. Celiac Rickets

IV. Hypo-phosphatasia

V. Hyper-parathyroidism

*Hey .. It's your way to
reach your aims*

Recent Classification

Type I: Iry calcium deficiency " Deficiency of $1,25 - (OH)_2 - D_3$ "

1. Nutritional deficiency.
2. Malabsorption of Vit-D
3. Hepatic disease
4. Toxic hydroxylation.
5. Renal Osteodystrophy
6. Vit-D dependent rickets type I

TYPE II: Iry Phosphate Deficiency : "No 2ry hyper-parathyroidism

1. Iry hypo-phosphatemic rickets.
2. De Toni-Fanconi syndrome
3. Lignac syndrome
4. Lowe's syndrome
5. Proximal renal tubular acidosis.
6. Phosphate deficiency or malabsorption.

TYPE III: End-Organ Resistance to $1,25 (OH)_2 - D_3$:

Vit-D – dependant rickets type II

TYPE IV: Related Conditions Resembling Rickets

- 1) Hypo-Phosphatasia
- 2) Hyper-Parathyroidism.

Vitamin - D

- It is a steroid, fat soluble vitamin.

Sources

1. Breast & Cow milk

⇒ both are deficient sources .

2. Rich sources are :

⇒ Cod liver oil, egg yolk (3-10 ug / gm) (1 ug = 40 IU),
fortified milks .

Requirements

The daily requirements are

- 400 – 800 IU/D for normal children.
- 800-1200 IU/D for PT & IUGR

Storage

⇒ Liver

Forms

1. Vit -D2 "Ergocalciferol"

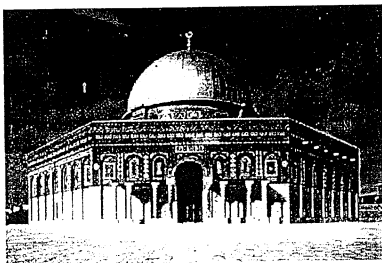
⇒ Produced by irradiation of plant "Ergosterol"

2. Vit-D3 "Cholecalciferol"

⇒ Produced by irradiation of 7-dehydro-cholesterol present in skin by UVR in the sun light (300–310 nm). These rays do not pass through ordinary window glass.

Metabolism

1. Absorption ⇒ from the small intestine along with fat in the presence of bile salts.
2. Both the absorbed Vit -D & that which is formed in the skin under the effect of UVR are carried on α 2 globulin to the liver.
3. In the liver it is hydroxylated to 25 (OH) cholecalciferol. Then carried to the kidneys to be hydroxylated at position "1" producing 1,25 dihydroxy cholecalciferol which is the active form of vit-D. If Ca & P is high, hydroxylation in 24 position to form less active form 24,25 (OH)₂-Vit-D3.



Come on .. Do your best

Vit - D Deficiency Rickets

Etiology

1. ↓↓ Intake (rickitogenic diet containing)

- a. Cow milk. ✓
- b. Delayed weaning. ✓
- c. low amounts of vit D & calcium. ✓
- d. High phytate or oxalate. ✓
- e. Bad Ca/P ratio. ✓
- f. Increased alkalinity (e.g. excess beverages) ✓
- g. ↓ Vit - C ✓

2. Insufficient exposure to sun ray "UVR"

- a. Glass windows & high buildings.
- b. Cloudy & foggy weather.
- c. Heavy pollution.
- d. Overlapped infant.
- e. Dark skin infant.

3. Malabsorption of Vit-D:

- a. Malabsorption syndrome.
- b. Obstructive jaundice

4. Deficiency of the active form:

- a. Vit-D dependent rickets type I
- b. Liver disease
- c. Toxic hepatic hydroxylation.

Contributing Factors

- 2. **Age:** 6 m - 2 years "due to rapid rate of growth"
- 3. **Rate of growth:** usually affecting preterm, & twins, but not those with slow arrested growth as crinism & marasmic patients.
- 4. **Race:** more common in dark races → Why?
- 5. **Season:** more in winter than in summer → Why?
- 6. **Atmosphere:** more common in big cities → Why?

Endochondral ossification

Normally, bone growth occurs by 2 types of ossification

1. Endochondral ossification

⇒ occurs in the epiphysis, leading to increased length of bone.

2. Subperiosteal ossification.

⇒ ↑↑ bone thickness.

In the normal epiphysis, the following layers are present & can be visualized microscopically:

a. Zone of resting cartilage

Composed of a single layer of non dividing resting cartilage cell

b. Zone of proliferating Cartilage "ZPC"

- Composed of 4-6 layers of dividing chondroblasts, regular in arrangement.

- It is not vascular & no calcium is deposited.

c. Zone of provisional calcification.

-The chondroblasts becomes swollen & vacuolated.

-The enzyme alkaline phosphatase splits PO₄ group from organic phosphate compounds in the local alkaline medium.

- This facilitate deposition of Ca⁺⁺ phosphate salts in the matrix forming the epiphyseal line.

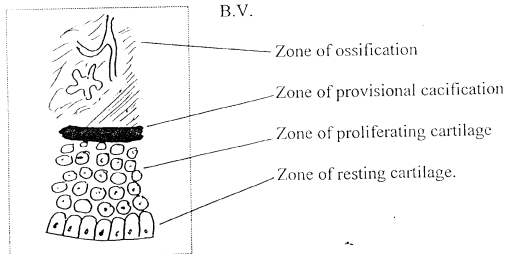
⇒ This line appears as sharp dense line at the end of metaphysis in X-ray.

d. Zone of ossification

- The degenerating cartilage cells are invaded by capillaries & connective tissue including osteoblasts, derived from B.M.

- The osteoblasts deposit a layer of osteoid which becomes rapidly mineralized.

So the cartilage is ultimately replaced by bone.



Pathology

In Active Rickets

1. There is failure of the cartilage to complete its cycle of proliferation & degeneration.
2. There's failure to deposit Ca^{++} salts $\Rightarrow$ excessive proliferation of cartilage.
3. This proliferating cartilage without calcification at the line of provisional calcification $\Rightarrow$ *Fraying* of this line in X-ray.
4. Failure of deposition of Ca^{++} salts at ZPC $\Rightarrow$ deposition of an uncalcified osteoid tissue.
5. When this tissue becomes compressed by pressure $\Rightarrow$ *Cupping & broadening* of the metaphysis occurs.
6. Calcium salts are resorbed from the pre-existing cortical bones due to physiological hyper-parathyroidism $\Rightarrow$ *Rarefaction* of bones in X-ray.
7. Sub-peri-osteal deposition of unmineralized osteoid tissue $\Rightarrow$ *Double periosteal lines* in X-ray.
8. The shaft loses its rigidity $\Rightarrow$ *deformity & fracture*.
9. The fractures are characteristically of *green-stick* type, which means that only one cortex of the bone is fractured while the other is intact.

In Healing Rickets

1. Calcification takes place in the ZPC. $\Rightarrow$ well demarcated line in X-ray film.
2. Its calcification is not continuous with the shaft i.e. uncalcified osteoid tissue between them.

In Healed rickets

1. The osteoid tissue between ZPC & the shaft becomes mineralized.
2. Through the process of bone remodeling the bone deformities are gradually corrected.
3. If deformities are severe $\Rightarrow$ Spontaneous correction may be impossible & surgery may be indicated.

C/P

C/O

1. Delayed motor milestones e.g. sitting, standing, crawling, walking....etc.
2. Delayed dentition.
3. Deformities of the bones.
4. One of the complications.

History:

History of

1. Inadequate Vit-D intake.
2. Lack of exposure to sun.
3. Cow milk feeding.
4. Anti-convulsant drugs.
5. Delayed weaning.
6. No supplementation of diet with Vit-D
7. Symptoms of complications.



Work hard

concentrate

O/E

A. Bony Manifestations

1. Skull

a. Craniotables

Which disappears at the age of 1 year even if untreated.

d. Delayed eruption of teeth.

3. Spine

a. Kyphosis.

b. Scoliosis.

c. Kypho-scoliosis.

4. Limbs

a. Broadening of the lower radio-ulnar epiphysis.

b. Marfan Sign ⇒ horizontal groove on both malleoli ⇒ due to hypertrophy of the epiphysis at two different sites.

c. Déformities.

Genu varum

Genu valgum

Genu recurvatum.

Coxa Vara

Bowing of the forearm

Contracted pelvis.

a. Delayed closure of A.F. " Anterior fontanell"

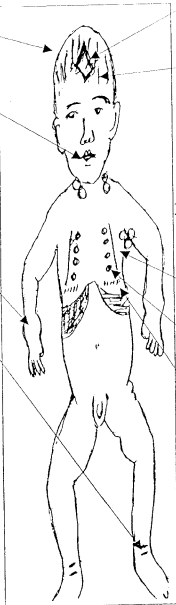
c. Frontal & parietal bossing ⇒ due to deposition of osteoid tissue at the center of ossification of these bones.

2. Chest

a. Pigeon Chest ⇒ The sternum is pushed forwards leaving a longitudinal groove on each sides.

b. Rickety rosary ⇒ thickening & broadening of the costo-chondral junction.

c. Harrison sulcus ⇒ horizontal groove at the lower part of the chest at the level of insertion of the diaphragm



5. Ligaments & Muscles

Laxity of ligaments, Hypotonia & Weakness of muscles ⇒

Kypho-scoliosis

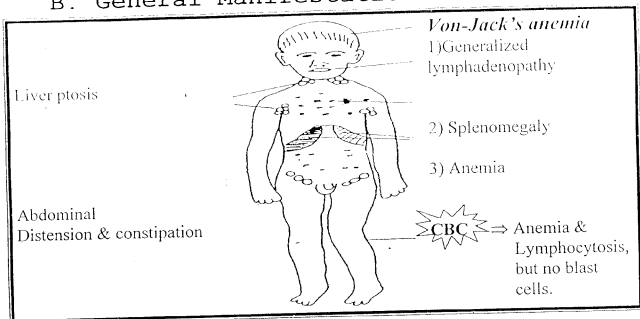
Pott belly abdomen

viscero-ptosis

Umbilical hernia.

It is due to hypo-phosphatemia.

B. General Manifestations.



Complications

1. Respiratory $\Rightarrow$ Pneumonia
Bronchopneumonia
Bronchitis
2. GIT $\Rightarrow$ Constipation due to hypotonia & GE.
3. Bone Fractures & deformities $\Rightarrow$ dwarfism
4. Anemia "Von Jack's anemia"
5. Tetany
6. Obstructed labor "later"
7. Dental caries

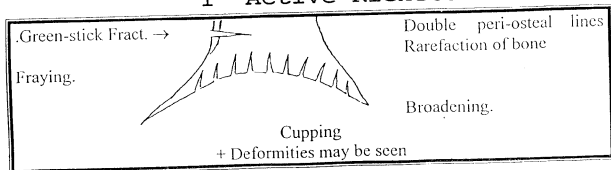
Q: What are the causes of tetany in Rickets?

1. Severe cases of rickets.
2. Failure of the parathyroid gland
3. Shock therapy with vit-D without calcium supplementation.
4. Bone stores of Ca^{++} are exhausted.

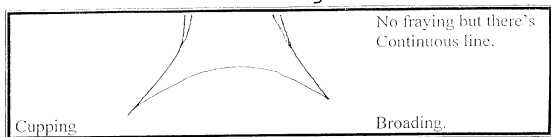
Investigations

A. X-ray of bones

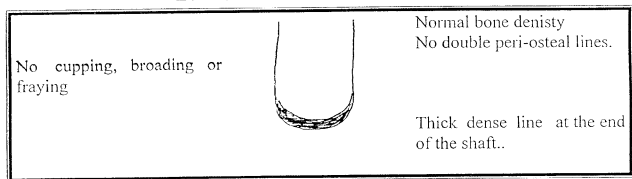
I- Active Rickets



II. Healing Rickets



III. Healed Rickets



B. Biochemical Changes

1. Serum Ca++ "N: 9-11 mg/dl"

⇒ Low normal

It may be reduced to tetanic level → Why?

N.B.

It is initially $\downarrow\downarrow \Rightarrow \uparrow\uparrow$ parathyroid hormone mobilization of Ca^{++}
From bones to the blood maintaining normal calcium level $\uparrow\uparrow$ Loss of
(P) in urine $\rightarrow \downarrow$ (P).

2. Serum Po_4 "N: 4.5 - 6.5 mg/dl"

→ Reduced 1.5-3 mg / dl

Ca X P (Howland formula) is low < 30 (N > 40).

3. Serum Alkaline phosphatase

→ Elevated > 500 IU / L "N: 145-200 IU / L" due to $\uparrow\uparrow$ Osteoblastic activity. It is 1st laboratory parameter to change in rickets & last to return to normal after complete healing.

Others

5. Parathyroid hormone level & urinary c-AMP $\Rightarrow \uparrow\uparrow\uparrow$.

6. Serum 25 (OH) D3 $\downarrow\downarrow$ (N: 20-80 ng / ml, being higher in summer) & serum 1,25 (OH)₂D3 is $\downarrow\downarrow$ (N: 25-45 pg / ml).

7. Generalized amino-aciduria: as vitamin D is essential for normal function of renal tubules.

8. Urine Ca⁺⁺ $\Rightarrow$ Absent.

9. Urine PO₄ $\Rightarrow \uparrow \uparrow \uparrow$

D.D.

I. From other causes of inability to walk.

II. From other causes of craniotabes:

1. Hydrocephalus.
2. Premature.
3. Congenital syphilis.
4. Osteogenesis imperfecta.
5. Some normal newborn near sutures.

III. Rickety rosary:

1. Hydrocephalus.
2. Critinism.

IV. Delayed closure of A.F.

1. Hydrocephalus
2. Critinism
3. Mongolism.
4. Osteogenesis imperfecta.

V. Delayed teething.

1. Critinism.
2. Mongolism.
3. Osteogenesis imperfecta.

VI. Spine deformity.

$\Rightarrow$ Pott's disease.

Other causes of short stature.

ttt

A. Prevention

1. Exposure to UVR
2. Supplementation of the diet with the daily requirements of vit. D, 400-800 IU/day.
3. Premature babies should receive 800-1200 IU/D as early as 2nd-4th month of life.

B. Active ttt

1. Vit-D:

2000-6000 IU/D for 2 months or shock therapy as 600,000 IU by deep IM/2 weeks for 3 doses.

2. Oral calcium intake together with vit-D

After complete healing $\Rightarrow$ Vit. D should be given in a dose
400- 800 IU/D

3. Surgical correction of deformities after complete healing.

4. ttt of complications.

Renal Rickets

Glomerular Ricket or Renal Osteodystrophy

Bone disease is one of the main features of chronic glomerular insufficiency & chronic renal failure.

Etiology

The most common causes are:

1. Chronic glomerulonephritis.
2. Chronic pyelonephritis.
3. Polycystic kidneys.
4. Chronic renal failure of any other causes.

Pathogenesis:

1. Renal parenchymal damage $\Rightarrow$ failure of renal hydroxylation of vit-D
2. $\downarrow\downarrow\downarrow$ GFR $\Rightarrow$ (P) retention $\Rightarrow \uparrow\uparrow$ (P)
 H^+ retention $\Rightarrow$ Acidosis.
 - a. $\uparrow\uparrow$ (P) $\Rightarrow \oplus$ Parathyroid gland $\Rightarrow \uparrow\uparrow$ PTH $\Rightarrow$ bone resorption & picture of osteitis fibrosa cystica.
 - b. Acidosis $\Rightarrow \uparrow\uparrow$ solubilization of bone Ca ++ salts.

C/P

1. Manifestations of CRF

- a. Marked growth failure & dwarfism due to
 - Under-nutrition.
 - Osteodystrophy
 - Hormonal abnormalities.
 - Medications esp. steroids.
- b. Anemia.
Due to
 1. $\downarrow\downarrow$ Erythropoietic factor
 2. Uremia $\Rightarrow \ominus$ BM
- c. Hypertension

2. Rickets

- ⇒ Bone deformities
- Pathological fracture.
- Dental abnormalities.

Investigations

A. Bone X-ray

- ⇒ Picture of rickets "active"
- Marked osteopenia & cystic like changes "Osteitis Fibrosa."
- Cystica → Why?

B. Biochemical

1. Ca++ ⇒ Normal or low
"No tetany due to acidosis"
2. (P) ⇒ ↑↑ "The only type of rickets in which (P) is high"
3. Alkaline phosphatase ⇒ ↑↑↑
4. Renal function tests ⇒ Impaired
⇒ ↑↑ Urea
↑↑ Creatinine.

C. CBC ⇒ Normocytic normochromic anemia.

t t t

1. t t t of the original renal condition

2. If there's Renal Failure

- a. Dialysis.
- b. Renal transplantation.
- c. Erythropoietin ⇒ for correction of anemia.
- d. NaHCO_3 ⇒ For correction of acidosis.

3. Rickets

- a. Restriction of (P) in diet & Oral administration of Aluminum hydroxide, Aluminum Carbonate ⇒ bind to (P) in intestinal lumen ⇒ ↓↓ absorption.
- b. ↑↑↑ Ca++ intake
- c. Active vit-D administration ⇒
e.g. Di-hydroxycholesterol "AT 10"
20 Ug/Kg/d
or Calcitriol "1,25 (OH)₂ - D₃"
20-50 ng/Kg/d
- d. Surgical correction of deformities
- e. Parathyroidectomy ⇒ if there's 2ry hyper-parathyroidism refractory to t t t.

Tubular Rickets

Primar hypophosphatemic rickets (XL-D)

⇒ Both males & females are affected, but the clinical picture is more sever in males.

Pathogenesis

2 Pathogenic mechanisms ⇒ Rickets

1. $\downarrow\downarrow$ Tubular reabsorption of (P) ⇒ (P) loss in urine "phosphaturia"
⇒ $\downarrow\downarrow$ (P)
In the blood "Hypo-phosphatemia":
2. Defective renal hydroxylation of vit-D at position 1

C/P

1. The general condition of the patient is good.
2. Waddling gait.
3. Short stature.
4. Teeth abnormalities.
5. Bowing of lower limbs, genu varum, genu valgum, coxa vara.
6. No tetany, rickety rosary or harrison sulcus.

Investigations

1. Serum Ca++ ⇒ Normal or slightly $\downarrow\downarrow$
2. Serum (P) ⇒ Markedly $\downarrow\downarrow\downarrow$
3. Alkaline phosphatase ⇒ $\uparrow\uparrow\uparrow$
4. No evidence of hyper-parathyroidism i.e. No cystic changes in the bones.

ttt

1. $\uparrow\uparrow\uparrow$ (P) intake:
Young children ⇒ 0.5 - 1 gm/day.
Older children ⇒ 1-4 gm/day
In the form of Na or K phosphate
2. Active Vit-D ⇒ as in Renal osteodystrophy.

De Toni Fanconi Syndrome

There's multiple defects of the proximal renal tubules to reabsorb the following:

1. (P) $\Rightarrow$ Phosphaturia + Hypophosphatemia
2. Glucose $\Rightarrow$ glucosuria.
3. A.A. $\Rightarrow$ generalized amino-aciduria. In addition, there may be:
4. K⁺ $\Rightarrow$ Hypokalemia + Hyperkaluria.
5. HCO₃ $\Rightarrow$ Bicarbonaturia + metabolic acidosis.

Etiology:

1. Idiopathic : is the most common:
It may be sporadic, AR or AD.
2. 2ry to: Renal toxin as mercury, lead, cadmium or exposure to outdated tetracyclines.
3. In association with certain inborn errors of galactosemia- Tyrosinemia- Lowe syndrome- Wilson disease.

C/P

1. Dwarfism + Failure to thrive.
 2. Rickets $\Rightarrow$ due to
 - a. Hypophosphatemia.
 - b. Acidosis.
- It doesn't respond to ordinary doses of Vit -D
3. The general condition is usually bad due to $\Rightarrow$ Acidosis.
 $\Rightarrow$ Hypokalemia.
 4. Vomiting, Polyuria, Polydipsia & Constipation.
 5. Weakness, fever with dehydration & metabolic acidosis.

Investigations

1. $\downarrow\downarrow$ K⁺
2. $\downarrow\downarrow$ P
3. Glycosuria.
4. Generalized aminoaciduria.

ttt

1. ttt of the cause in 2ry type.
2. Active vit. D : as renal osteodystrophy.
3. $\uparrow\uparrow\uparrow$ (P) intake : as before.

4. Adequate calcium intake.
5. ↑↑↑ (K) intake : KCL 2-3 mEq/Kg/D.
6. NaHCO₃ : For correction of acidosis.

Lignac syndrome (AR)

There is accumulation of cystine in various tissues (spleen , liver , BM, LN ,kidney ,cornea & conjunctiva)

C/P

As Fanconi \$ in addition to :

1. Photophobia due to corneal affection.
2. HSM.
3. Renal failure may occur.

Investigations:

1. Slit lamp examination of cornea.
⇒ refractile cystine crystals.
2. Lab. Findings of fanconi syndrome.

ttt

As fanconi syndrome +

1. D-Penicillamine ⇒ for chelation of cystine.
2. Recently: Cysteamine ⇒ binds to SH-group of cystine.
3. If RF ⇒ Dialysis or renal transplantation.

Infantile Renal Tubular Acidosis: Lightwood \$ "AR"

RTA is classified into

Proximal RTA (type II) : ⇒ Resulting from reduced proximal tubular reabsorption of HCO₃.

Distal RTA (type I) : ⇒ Resulting from reduced distal tubular secretion of hydrogen ion..

1. Metabolic acidosis.
2. Hypophosphatemia.
3. Hypokalemia.
4. Hyperchloremia.

ttt

1. ttt of acidosis & hypokalemia.
2. Correction of hypophosphatemia.
3. Active form of Vit-D

Lowe's "Oculo-Cerebro-Renal (XLR)

1. Ocular $\Rightarrow$ Cong cataracts-Glucoma Buphthalmos.
 2. Cerebral $\Rightarrow$ MR- severe hypotonia & hypo-reflexia.
 3. Renal $\Rightarrow$ Inability to acidify urine & features of fanconi syndrome.
- Rickets is due to acidosis & hypophosphatemia.

Vit.D Dependent Rickets "AR"

Type I

$\Rightarrow$ Due to hereditary defect in renal hydroxylation of Vit. D $\Rightarrow \downarrow \downarrow$
Ca $^{++}$ & P $+$ $\uparrow \uparrow$ Alkaline phosphatase $\Rightarrow \uparrow \uparrow$ PTH

ttt

Active form of Vit. D

Type II

$\Rightarrow$ Due to target organ resistance to Vit. D
There is short stature & alopecia totalis.
Rickets may respond to very high doses of Vit D "active form"



To be the first..
You must have big harvest

Hepatic Rickets

Due to

1. Defective hydroxylation at position 25.
2. Malabsorption of Vit-D due to absence of bile salts in cases of obstructive Jaundice.
3. Toxic hepatic hydroxylation - in case of anti-convulsant therapy.

These drugs accelerate metabolism & catabolism of Vit-D.

Celiac rickets

Due to malabsorption of Vit-D "Fat soluble" $\Rightarrow$ These F.A. will bind to $\text{Ca}^{++} \Rightarrow \text{Ca}^{++}$ soaps formation $\Rightarrow$ will precipitate in the intestine $\Rightarrow \text{Ca}^{++}$ malabsorption.

Hypo-Phosphatasia AR

- Due to cong. Absence or deficiency of alkaline phosphatase $\Rightarrow$ defective mineralization of bones.
- Serum Ca^{++} is high $\Rightarrow$ Nephro-calcinosis
- Seru. ALK phosph $\Rightarrow$ Markedly low.
- Urine characteristically contains increased amount of phospho-ethanolamine.

!!!

$\Rightarrow$ infusion of plasma rich in Alkaline phosphatase.

Hyper-parathyroidism

Hyperplasia, adenoma or carcinoma of the parathyroid gland $\Rightarrow \uparrow\uparrow$ PTH $\Rightarrow$ picture of osteitis fibrosa cystica. Both serum Ca^{++} & alkaline phosphatase are increased.

!!!

Adenoma $\Rightarrow$ excision

Hyperplasia $\Rightarrow$ total parathyroidectomy + post operative supplementation with Ca^{++} is essential.

Nutritional Assessment

It is important for prevention, early detection & ttt of nutritional disorders.

Methods

1. Dietary evaluation.
2. Clinical evaluation.
3. Anthropometric evaluation.
4. Laboratory findings.

I. Dietary evaluation

1. Measuring the quantity & quality of food intake regarding the major food elements, vitamins, minerals and trace elements & comparison with the individuals requirements.
2. History of any abnormality in feeding practice.
3. Collection of information about food consumption by:
 - a. Records based on 24 hrs recall
 - b. 3-7 days feeding history or,
 - c. The usual pattern of food consumption. "quantity& quality"

II. Clinical evaluation

By full clinical examination we can detect cases of severe nutritional deprivation or even mild to moderate nutritional disorders.

III. Anthropometric evaluation

It is the measurement of physical dimensions of the human body at different ages & comparison with standard references. To determine abnormalities in growth.

1. Weight "Wt"

- $\Rightarrow$ Good index for acute nutritional status.
- It is plotted on 2 graphs $\Rightarrow$ Wt. for age.
Wt for height.
- Under - weight is less than the 10th percentile.
- Over - weight is more than the 90th percentile.

2. Length ~ Ht

- $\Rightarrow$ It indicate chronic growth failure which may be due to chronic under nutrition
- If $< 10^{\text{th}}$ percentile $\Rightarrow$ Dwarfism.

3. Wt for Ht

- It is important for measuring body built & distinguishes wasting from dwarfism.

4. Growth Velocity:

- It is expressed by cm / Year & plotted on special curves.
- It is a more sensitive method for assessing growth failure or slow growth.

5. Triceps skin fold thickness.

$\Rightarrow$ An indicator for total body fat.

Because about $\frac{1}{2}$ body fat is in the S.C. layer.

It is measured at the midpoint between the acromion & olecranon at the non dominant arm during relaxation.

It is more than 13mm after 1 year age.

6. Mid-arm circumference & ms. mass.

By measuring the triceps skin fold thickness & the midarm circumference, we can calculate the ms. mass, which is a good indicator of nutritional status.

The mid-arm circumference is normally more than 13.5 cm after 1 year of age.

7. Chest / head circumference ratio

- At birth this ratio is less than 1
 - At 6 months $\Rightarrow$ "1" & gradually increases later on.
- A ratio < 1 after 6m is one of the early features of malnutrition.

IV. Laboratory Evaluation

It is used to confirm nutrient deficiencies which has been determined by dietary, clinical or anthropometric evaluation.

It can also help in diagnoses of subclinical cases e.g.

Hb %, Hct, S. albumin & other proteins ... etc.

It almost been done

Vitamins & Minderals Deficiency

Vit - B2

1. Angular stomatitis
2. Glossitis
3. Vascularization of the Cornea.

ttt

⇒ 10 mg/ D oral
+ other Vit.

Nicotinamide

1. Smooth red tongue
2. 4 Ds ⇒ Dementia.
Diarrhea
Dermatitis
Death

ttt

⇒ 50-300 mg/D oral or IM
+ well balanced diet.

Vit-C

⇒ Scurvy

1. Hgic manifestations "gums"
2. Poor wound healing.
3. Sub-periosteal hge.
4. Tender limbs.
5. Susceptibility to infection

ttt

⇒ 100 -200 mg/D oral or IV
or orange juices.

Vit-A

Eyes ⇒ Night blindness
Bitot spots
Xerosis
Corneal ulceration

Skin $\Rightarrow$ Hyperkeratosis follicularis "Goose skin"

!!!

5000 U/D oral + 25.000 U/M till recovery.
Then 5000 U/D (Oral)

Vit-K

Bleeding tendency due to hypoprothrombinemia "II, VII, IX, X"

!!!

1 mg/Kg/D

Vit-E

Hemolytic anemia in premature

Vit-B1

$\Rightarrow$ Beri beri

Vit-B6

CNS $\Rightarrow$ convulsions + polyneuropathy
Anemia $\Rightarrow$ Hypochromic
Glossitis, seborrhea.

Copper

$\Rightarrow$ Anemia + hair changes

Zinc

$\Rightarrow$ Acrodermatitis enteropathica.

Vitamin Toxicity or Hypervit.

Hypervitaminosis D

Usually due to high doses > 20.000 U/D for 1-3 m.

C/P

1. GIT: Anorexia
Vomiting
Constipation
2. Kidney: Polyuria

Dehydration
Renal stones
Nephrocalcinosis.

3. Generalized weakness, hypotonia.

4. Hypercalcemia $\Rightarrow$ Hypercalcuria
Metastatic calcifications "Renal & other tissues"
Calcific aortic stenosis

Investigations

1. Serum $\text{Ca}^{2+} \Rightarrow \uparrow\uparrow\uparrow$
2. $\uparrow\uparrow\uparrow \text{Ca}^{2+}$ in urine "Hypercalcuria"
3. X-ray $\Rightarrow$ show metastatic calcification
Nephrocalcinosis
Osteoporosis.

D.D.

1. Hyperparathyroidism
2. Idiopathic hypercalcemia
3. Chronic renal failure.

ttt

A. Prevention

$\Rightarrow$ Avoid high doses of Vit.D. for long periods.

But if this is necessary, monitoring of serum Ca^{++} is important.

B. Curative

1. Stop Vit-D till symptoms disappear & serum Ca^{++} becomes normal
2. Low Ca^{2+} diet.
3. Correction of dehydration.
4. $\text{Al}(\text{OH})_3$ gel orally $\Rightarrow$ to $\downarrow\downarrow$ Ca^{2+} absorption.
5. EDTA $\Rightarrow$ to chelate Ca^{++}
6. Cortisone in severe cases
 $\Rightarrow$ Why?

Hypervitaminosis A

A. Acute

⇒ usually after ingestion of 300,000 U or more.

C/P ⇒ Nausea, vomiting + ↑ ICP ⇒

Bulging fontanel, papilledema,

⇒ Pseudo-tumor cerebri

B. Chronic

Anorexia

Tender swelling of bones.

Cranioabomas

Desquamation of palms & soles.

Fissuring of angles of mouth.

X-ray ⇒ cortical hyperostosis of bones.

Vit- K toxicity

Synthetic vit-K is an oxidizing agent

⇒ hemolysis of RBCs ⇒ Hyper-bilirubinemia & kernicterus in

The premature & G-6 - P D deficient newborn.

Infantile Tetany

It is a state of hyper-excitability of the central & peripheral nervous system resulting from abnormal ionic concentration in the fluid bathing nerve cells & peripheral nerves.

Etiology

A. Hypocalcemia

1. Vit - D deficiency or abnormalities in Vit-D metabolism.

2. Hypo-parathyroidism.

a. Early neonatal hypocalcemia (1st 72 hrs) ⇒ infant of diabetic mother, preterms, IUGR, perinatal asphyxia.

b. Late neonatal hypocalcemia (5-10 days) ⇒ A high phosphate food such as cow milk depresses serum Ca through the deposition of Ca in bone while the infant parathyroid is not yet able to respond by ↑↑ PTH.

- c. Aplasia or hypoplasia of parathyroid e.g. DiGeorge \$, CHARGE or VATER \$.
 - d. Surgical removal parathyroid gland.
 - e. Infiltration of parathyroid gland as in hemosiderosis or Wilson's disease.
 - f. Autoimmune destruction of parathyroid gland.
 - g. Familial.
3. Exchange transfusion.

B. Alkalosis.

1. Hyper-ventilation.
2. Alkali-therapy " NaHCO_3 "
3. Excessive loss of gastric HCL e.g. "Vomiting"

C. Hypomagnesemia.

1. Chronic diarrhea.
2. Malabsorption of Mg^{++}
3. Exchange transfusion.
4. Total parenteral nutrition without sufficient Mg.
5. Nephrotoxic medication & diuretic therapy.

C/P

A. Manifest tetany

If serum $\text{Ca}^{++} < 7 \text{ mg/dl}$

1. Carpo-pedal spasm
2. Laryngeal spasm
3. Convulsions in infants & newborn.

B. Latent Tetany.

If serum Ca^{2+} is $7-9 \text{ mg/dl}$

1. Chevostek sign

Tapping the facial nerve "anterior to external auditory meatus" $\Rightarrow$ contraction of orbicularis oris ms.

2. Trousseau sign

Occluding the arterial flow to the forearm by inflating a blood pressure cuff over systolic pressure for 3 min $\Rightarrow$ appearance of the typical carpo-pedal spasm.

3. Peroneal sign

Tapping the peroneal nerve over the neck of the fibula $\Rightarrow$ dorsi-flexion & eversion of the foot.

4. Prolonged QT interval.



ttt

A. Immediate

1. Ca gluconate 10 %

⇒ 2ml / Kg slowly IV

2. If no response ⇒ search for the etiology & correct the alkalosis or Mg deficiency "0.2 ml/kg IM"

B. Maintenance

1. Diet rich in Ca^{++} or Ca chloride orally.

2. Vit – D for ttt of rickets & improving Ca-absorption.



You need help..
We're here